Guillain-Barré Syndrome

Section 1: ABOUT THE DISEASE

A. Etiologic Agent

Guillain-Barré syndrome (GBS) is a rare, autoimmune disorder that affects the peripheral nervous system. GBS is characterized by temporary paralysis, usually beginning in the lower extremities and moving to the upper extremities, with diminished to lost reflexes. In about 66% of cases, an antecedent infection (occurring in the days to weeks preceding GBS symptom onset) is believed to trigger an immune response that reacts with nerves. Respiratory and gastrointestinal infections are the most commonly reported antecedent events. The most commonly reported infection preceding GBS is infection with *Campylobacter jejuni* (see chapter titled *Campylobacter Enteritis* for more information). In recent studies, 26–36% of GBS patients have serologic or culture evidence of preceding *C. jejuni* infection. Risk of GBS is also associated with pre-existing cytomegalovirus (CMV) infection and genetic predisposition.

Occasionally, immunization is thought to trigger the syndrome. While associations have been proposed between GBS and several vaccines, including rabies, oral polio, influenza, measles/mumps/rubella (MMR), measles, tetanus toxoid-containing vaccines, and hepatitis B, a cause-effect relationship between GBS and these vaccines has not been clearly identified. There is an increased risk of GBS associated with vaccines made in neural tissue, such as rabies vaccine produced in suckling mouse brain tissue, but such vaccines are not licensed for use in the U.S. There was an association of a higher than expected rate of GBS in individuals who received an influenza vaccine used for a potential pandemic strain of influenza A in 1976, but no association has been demonstrated with any other influenza vaccine.

After a review of data pertaining to influenza vaccine, the Advisory Committee on Immunization Practices (ACIP) concluded in its 1999 recommendations that:
"Even if GBS were a true side effect of vaccination in the years after 1976, the estimated risk of GBS of slightly more than one additional case per million persons vaccinated is substantially less than the risk for severe influenza, which could be prevented by vaccination in all age groups…The potential benefits of influenza vaccination in preventing serious illness, hospitalization, and death greatly outweigh the possible risks for developing vaccine-associated GBS."

In more recent ACIP statements, the ACIP reiterates that no evidence exists for a cause-effect relationship between influenza vaccines and GBS.

B. Clinical Description

The first symptoms of GBS are varying degrees of weakness or tingling sensations in the legs and weakening of the anal sphincter. In many cases, the weakness spreads to the upper body. The symptoms increase in intensity until the patient may become totally paralyzed. When this happens, paralysis interferes with respiration, requiring that the patient be placed on a ventilator. Complications can include infection, blood clots, and low or elevated blood

pressure. After the onset of GBS, symptoms typically progress over the course of hours, days, or weeks. The stage of greatest weakness usually occurs between 2–4 weeks after onset. Most patients recover completely from even the most severe cases of GBS. However, some patients experience residual weakness.

C. Vectors and Reservoirs

C. jejuni, a bacterium that appears to cause the most commonly reported infection preceding GBS, is widely prevalent in the gastrointestinal tracts of many animals, notably cattle and poultry, swine, sheep, and even in pets such as birds, kittens, and puppies. Raw poultry is commonly contaminated with *C. jejuni*.

D. Modes of Transmission

While GBS itself is not communicable from person to person, infections that precede GBS may be infectious.

E. Incubation Period

Since, in most cases, the trigger of GBS is not known, the incubation period cannot be determined. If a preceding respiratory or gastrointestinal illness is identified, it typically occurs 1–3 weeks prior to the onset of GBS symptoms.

F. Period of Communicability or Infectious Period

While GBS itself is not communicable from person to person, people may still shed infectious agents from the infection that preceded GBS.

G. Epidemiology

GBS is rare, affecting only about 1/100,000 persons each year, with males and females affected equally. GBS can occur at any age but appears to be more common in young adults and the elderly. There is no well-defined geographic distribution.

H. Bioterrorist Potential

Not applicable.

Section 2:
REPORTING CRITERIA AND LABORATORY TESTING

A. What to Report to the Massachusetts Department of Public Health (MDPH)

Report any clinically diagnosed case of GBS, as reported by a health care provider.

B. Laboratory Testing Services Available

Testing services for an infectious agent that may have triggered GBS may be available through the MDPH State Laboratory Institute (SLI). Consult with the epidemiologist on-call at the MDPH Division of Epidemiology and Immunization at (617) 983-6800 or (888) 658-2850 for more information on testing.

Section 3:

REPORTING RESPONSIBILITIES AND CASE INVESTIGATION

A. Purpose of Surveillance and Reporting

- To further assess the burden of disease caused by *C. jejuni* infections.
- To monitor GBS cases for associations with infectious diseases.
- To collect data and to address concerns relating to GBS and vaccine safety.

B. Laboratory and Health Care Provider Reporting Requirements

GBS is reportable to the local board of health (LBOH). The MDPH requests that health care providers immediately report to the LBOH in the community where the case is diagnosed, all confirmed or suspect cases of GBS, as defined by the reporting criteria in Section 2A.

C. Local Board of Health (LBOH) Reporting and Follow-up Responsibilities

Reporting Requirements

MDPH regulations *(105 CMR 300.000)* stipulate that GBS is reportable to the LBOH and that each LBOH must report any confirmed case of GBS or suspect case of GBS, as defined by the reporting criteria in Section 2A. Cases should be reported to the MDPH Bureau of Communicable Disease Control, Office of Integrated Surveillance and Informatics Services (ISIS) using a MDPH *Guillain-Barré Syndrome Case Report Form* (found at the end of this chapter). Refer to the *Local Board of Health Timeline* at the end of this manual's *Introduction* section for information on prioritization and timeliness requirements of reporting and case investigation.

Case Investigation

1. It is the responsibility of the LBOH to complete a MDPH *Guillain-Barré Syndrome Case Report Form* by interviewing the case and others who may be able to provide pertinent information. Much of the information on the form can be obtained from the health care provider or from the medical record.
2. Use the following guidelines to assist in completing the form:
   a. Accurately record the demographic information.
   b. Record all available clinical information, including date of symptom onset, symptoms, clinician contact information, and hospitalization dates and location(s).
   c. Record the history of illness, including whether or not the case had fever, respiratory illness, gastrointestinal illness, or immunizations within the eight weeks preceding GBS.
   d. If you have made several attempts to obtain case information but have been unsuccessful (e.g., the case or health care provider does not return your calls or respond to a letter, or the case refuses to divulge information or is too ill to be interviewed), please fill out the case report form with as much information as you have gathered. Please note on the form the reason(s) why it could not be filled out completely.

3. After completing the case report form, attach laboratory report(s) and fax or mail (in an envelope marked "Confidential") to ISIS. The confidential fax number is (617) 983-6813. Call ISIS at (617) 983-6801 to confirm receipt of your fax. The mailing address is:

MDPH, Office of Integrated Surveillance and Informatics Services (ISIS)
305 South Street, 5th Floor
Jamaica Plain, MA 02130
Fax: (617) 983-6813

4. Institution of disease control measures is an integral part of case investigation. It is the responsibility of the LBOH to understand, and if necessary, institute the control guidelines listed in Section 4.

Section 4:
CONTROLLING FURTHER SPREAD

A. Isolation and Quarantine Requirements *(105 CMR 300.200)*

None.

B. Protection of Contacts of a Case

None.

C. Managing Special Situations

Reported Incidence Is Higher Than Usual

If the number of reported cases of GBS in your city/town is higher than usual, consult with the epidemiologist on-call at the MDPH Division of Epidemiology and Immunization at (617) 983-6800 or (888) 658-2850. The Division can help determine a course of action to prevent further cases and can perform surveillance for cases across town lines, which would otherwise be difficult to identify at the local level.

D. Preventive Measures

None.

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FORMS & WORKSHEETS

Guillain-Barré Syndrome

Guillain-Barré Syndrome

LBOH Action Steps

This form does not need to be submitted to the MDPH with the case report form. It is for LBOH use and is meant as a quick-reference guide to Guillain-Barré syndrome case investigation activities.

LBOH staff should follow these steps when Guillain-Barré syndrome (GBS) is suspected or confirmed in the community. For more detailed information, including disease epidemiology, reporting, case investigation, and follow-up, refer to the preceding chapter.

- ❑ Notify the MDPH Division of Epidemiology and Immunization at (617) 983-6800 or (888) 658-2850 to report any suspect or confirmed case(s) of GBS.
- ❑ Fill out the case report form (attach laboratory results).
- ❑ Send the completed case report form (with laboratory results) to the MDPH Bureau of Communicable Disease Control, Office of Integrated Surveillance and Informatics Services (ISIS).